

Case report

Eosinophilic fasciitis presenting as psoriatic arthropathy

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We describe a patient with eosinophilic fasciitis, a rare scleroderma-like steroid-responsive condition. Initial presentation suggested pauciarticular psoriatic arthritis.

CASE HISTORY. A 19-year-old student nurse presented with a four month history of pain and stiffness in her knees, ankles and elbows following a brief 'flu-like illness. Wrist and elbow movements were limited, with bilateral knee effusions and flexion deformities of approximately 10°. Evidence of psoriasis was limited to areas of hyperkeratosis on the elbows, and a few nail pits; there was a positive history in her maternal grandmother. Rheumatoid factor test was negative, serum C-reactive protein 19.5 mg/l (normal < 6), and erythrocyte sedimentation rate 39 mm/hr. X-rays of hands, elbows and knees were normal. A provisional diagnosis of psoriatic arthropathy was made and she was treated with the nonsteroidal anti-inflammatory drug nabumetone 1 gm daily, with improvement.

Five months later her condition deteriorated, her skin felt "very tight", and she had to walk on her toes because of tightness at the ankle. She described occasional difficulty with swallowing but gave no history of Raynaud's phenomenon. She now had thickened, shiny skin on the limbs and flexion deformity of her knees, wrists, and the small joints in her hands. Systemic sclerosis was considered to be the likely diagnosis, and she was commenced on d-penicillamine 100 mg/day and nifedipine 10 mg/tid.

Investigations showed eosinophilia of 8%; ESR 32 mm; C-reactive protein < 6 mg/l; serum IgM 2.72 g/l. Relevant autoantibody tests were negative (including antinuclear factor, anti-centromere antibody, anti SCL-70 and anti-neutrophil cytoplasmic antibody); complement components were normal and

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circulating immune complexes were not detected (Clq binding). Isotope bone scan revealed synovitis in both wrists, the metacarpophalangeal joints of both hands and ankles. *Borrelia burgdorferi* titres were negative. Barium swallow was normal.

Full thickness skin biopsy showed an intact epidermis, without hyperkeratosis or follicular plugging, and there was no vacuolar degeneration of the basal layer. There was loss of hair follicles, and the skin appendage structures were small and atrophic. The papillary dermis and upper reticular dermis were relatively normal but the deeper dermis showed thickened hyalinized eosinophilic collagen fibres (Figure). The dermis appeared thickened and there was extension of the collagen into the underlying subcutis. The fibrous septa within the subcutaneous fat were widened and within the fibrous tissue there were scattered chronic inflammatory cells at the periphery of the lobules of fat. The infiltrate was composed predominantly of lymphocytes and plasma cells with scattered histiocytes, but no eosinophils. The fascia was thickened, homogeneous and eosinophilic and was permeated by a chronic inflammatory cell infiltrate. A little underlying skeletal muscle was present in the biopsy and this showed myofibril degeneration and interstitial fibrosis. The overall appearances were those of eosinophilic fasciitis (Shulman's syndrome). Following histological diagnosis she was commenced on hydroxychloroquine sulphate 200 mg/day, prednisolone 40 mg/day and cimetidine 400 mg/twice daily; d-penicillamine was discontinued. She made a slow but gradual improvement, with return to normal gait.

It has been necessary to continue steroids at a relatively high dose.

DISCUSSION

Over 200 cases of eosinophilic fasciitis have been described since the initial description by Shulman.¹ It is a scleroderma-like disease characterised by symmetrical widespread inflammation and sclerosis of the deep fascia, subcutis and dermis: it has been termed *morphoea profunda* by some authors. It primarily involves the extremities, and is associated with peripheral blood and tissue eosinophilia, and often hypergammaglobulinaemia. Although well described, onset with polyarthritides may lead to initial diagnostic difficulty. Synovitis of large joints, hyperkeratosis and nail pits in conjunction with a family history of psoriasis, is a common mode of presentation of the pauciarticular type of psoriatic arthritis, and we believe this is the first report of eosinophilic fasciitis presenting in this manner. Synovitis had been present for at least four months prior to the insidious onset of sclerotic skin changes.

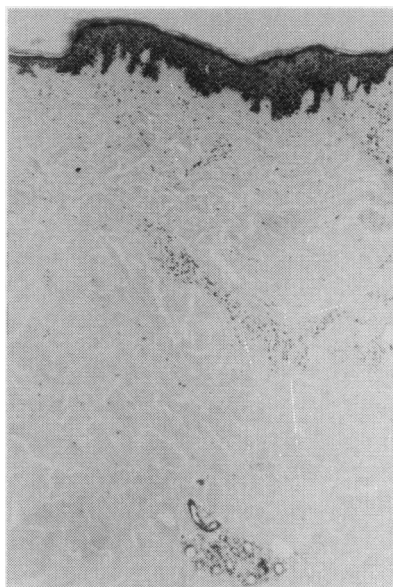


Figure. Thickened dermis and hyalinization of collagen bundles with chronic inflammatory cell infiltrate.

Eosinophilic fasciitis may be part of the spectrum of systemic sclerosis, as after one or two years a proportion of patients develop chronic fibrotic cutaneous features and some develop systemic manifestations, but there are some significant differences. These include the relative absence of Raynaud's phenomenon, normal nailfold capillary microscopy,² sparing of the epidermis and dermis, infrequent visceral involvement, absence of the serological features which characterise systemic sclerosis and the rare development of haematological complications.³ The male to female ratio is approximately equal, and in a recent review of 52 patients at the Mayo Clinic⁴ the mean age of onset was 47 years. In 46% of these cases, onset was related to unaccustomed strenuous exertion. Some authors mention a prodromal febrile illness as described by our patient, perhaps suggesting a viral trigger factor.⁵ Cutaneous manifestations are usual presenting features, and evolve through three stages: pitting oedema at onset, *peau d'orange* or dimpling, and finally induration. The most common sites are the arms and legs, with infrequent involvement of hands and feet. Isolated cases of oesophageal, cardiac or pulmonary disease have been described, but in contrast to systemic sclerosis these are uncommon features. In over 60% of cases eosinophilia is greater than 7% of the total white cell count, although peripheral eosinophilia can be transient, even in the absence of corticosteroid treatment. In our patient, eosinophilia was detected only once in her year-long illness.

To confirm the diagnosis a deep skin biopsy, including tissue from the epidermis down to the skeletal muscle through the deep fascia, must be performed. Changes range from inflammation with minimal connective tissue change to severe sclerosis. The deep fascia and septa of the subdermal fat are most extensively involved while the epidermis is normal or only slightly atrophic. Despite the name, the presence of eosinophils in the inflammatory infiltrate is not required for diagnosis. It is essentially a corticosteroid-responsive benign condition but patients with chronic ongoing disease have been described. In the larger proportion of cases resolution, which may be spontaneous, occurs within five years. Relapses, although unusual, do occur. Our patient displayed a characteristic clinical response to prednisolone. Cimetidine has also been reported to be effective,^{6,7} perhaps by involving the capacity of H₂ receptor antagonists to interfere with suppressor T-cell control.⁸

The etiology of eosinophilic fasciitis is unknown. One report of the condition developing in siblings not recently in contact within six months suggests the influence of genetic factors: they shared HLA A,B,C,DR, and DQ antigens.⁹ Hypersensitivity reactions to muscle tissue following exercise-induced damage and an association with cancer have also been demonstrated. An unusual syndrome characterised by incapacitating myalgia and peripheral eosinophilia in patients taking L-tryptophan has recently been reported. Some of these patients developed sclerotic cutaneous changes and the condition has been termed the eosinophilia myalgia syndrome. Overlapping clinical features suggested a relationship of both these syndromes with the toxic oil syndrome, a multi-system disease associated with ingestion of adulterated cooking oil in Spain in 1981. Although these three conditions display many features in common, Shulman¹⁰ described distinguishing clinical and laboratory features. Polyarthritides appeared to be equally prevalent in all three conditions. The recent suggestion that *Borrelia*

burgdorferi infections may be responsible for scleroderma variants including eosinophilic fasciitis remains speculative.¹¹

This case illustrates the need for precise diagnosis in patients who present with inflammatory polyarthritis and cutaneous sclerosis. Systemic sclerosis is unresponsive to steroids and the prognosis is often poor. In contrast, patients with eosinophilic fasciitis respond well to steroids or other potent immunosuppressive therapy and complete remission may occur. Appropriate full-thickness skin biopsy must be considered in all patients with clinically atypical systemic sclerosis.

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